



Preoperative endoscopic extraction of a huge pancreatic protein plug in a choledochal cyst: A case report



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ABSTRACT

A 4-year-old female was referred to our institution with acute pancreatitis due to a choledochal cyst. A protein plug was found to be deeply impacted in the main pancreatic duct. She underwent preoperative endoscopic retrograde cholangiopancreatography (ERCP) followed by sphincteroplasty and extraction of the protein plug with a basket extractor. After the ERCP procedure, the patient successfully underwent total choledochal cyst excision with Roux-en-Y hepaticojejunostomy. In cases in which a huge protein plug is located deep in the pancreatic duct, therapeutic ERCP is useful as preoperative management for choledochal cyst excision with hepaticojejunostomy.

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An intraductal pancreatic protein plug is a worrisome problem in patients with a choledochal cyst. However, its surgical treatment has not been well discussed. We describe a 4-year-old female with a choledochal cyst that was accompanied by a huge protein plug that was impacted in the distal main pancreatic duct, and which was extracted endoscopically.

1. Case report

A 4-year-old female presented with abdominal pain and vomiting, and was diagnosed with a choledochal cyst by abdominal ultrasound and acute pancreatitis based on elevated serum pancreatic enzymes (amylase 216 IU/L, lipase 323 IU/L) and LFTs (TBil 1.55 mg/dL, DBil 0.64 mg/dL, AST 260 IU/L, ALT 499 IU/L, GGTP 761 IU/L).

Abdominal ultrasound, CT and MR-cholangiopancreatography (MRCP) revealed cystic dilation of the common bile duct (maximum diameter 17 mm) accompanied by a slightly dilated intrahepatic duct. The choledochal cyst was equivalent to Todani classification IV-A. Furthermore, a radiolucent filling defect (2 cm in length) was

detected in the main dilated pancreatic duct to the common channel by MRCP and abdominal ultrasound. While her symptoms of pancreatitis resolved by conservative therapies within one week after admission, the pancreatic substance did not disappear and remained lodged in the main pancreatic duct.

We performed diagnostic and therapeutic endoscopic retrograde cholangiopancreatography (ERCP) under general anesthesia preoperatively. The scope used for this patient was manufactured for adult use (Olympus JF-240, Olympus, Ltd., Tokyo, Japan: scope diameter 12.6 mm with a 3.2 mm channel). In ERCP, the common bile duct was expanded and the patient had a long common channel (30 mm in length) with a pancreaticobiliary junction. The pancreatic intraductal filling defect was a huge protein plug (20 mm × 9 mm) that was located in the main pancreatic (Wirsung's) duct (Fig. 1). The substance was unmovable despite the washout-effect of the contrast agent. Therefore, endoscopic extraction was performed following sphincteroplasty. A guide wire was placed in the pancreatic duct and a balloon dilator (6 mm in diameter) was inserted into the duct over the wire. After sphincteroplasty, the substance was retrieved with a basket lithotripter as a mass via the papilla of Vater and it was proved to be a protein plug (Fig. 2). Complete clearance was confirmed by cholangiography.

She did not develop post-ERCP pancreatitis and underwent surgical treatment consisting of excision of the choledochal cyst and Roux-en-Y hepaticojejunostomy. The collected gallbladder bile demonstrated elevated amylase (41,236 IU/L), which was due to the

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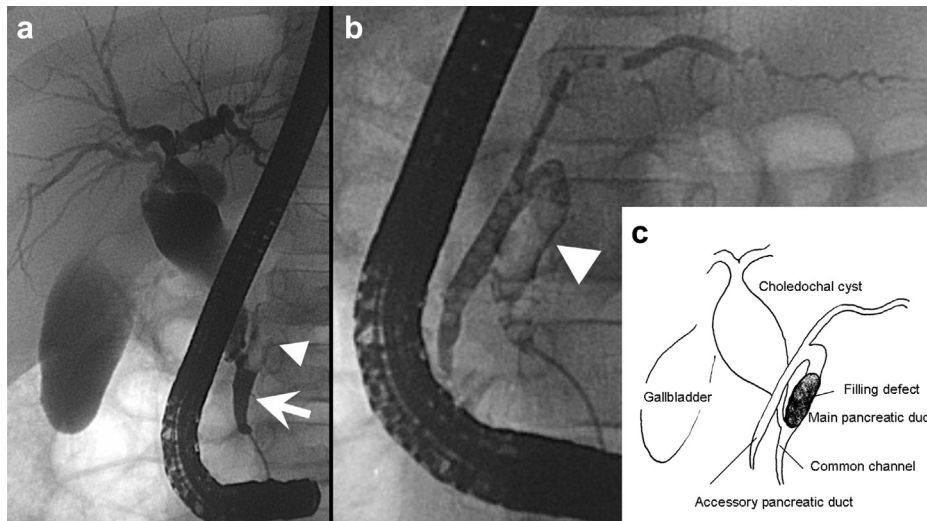


Fig. 1. ERCP showing a choledochal cyst with a pancreaticobiliary maljunction complicated with a huge obstructing protein plug in the main pancreatic duct. a) cholangiography, b) pancreatography, c) gross scheme. ERCP shows a pancreaticobiliary maljunction forming the common channel (arrow) for a length of up to 30 mm. The pancreatic protein plug was detected as a huge filling defect (arrowhead: 20 mm × 9 mm), which impacted and caused dilatation of the main pancreatic (Wirsung's) duct.

reflux of pancreatic enzymes into the common duct. The post-operative course was uneventful.

2. Discussion

In the common form of choledochal cyst, a pancreaticobiliary maljunction permits the reflux of pancreatic enzymes into the common bile duct through the maljunction, and it has been proposed that the associated symptoms are caused by this reflux [1]. On the other hand, a protein plug may also be found in the common channel or pancreatic duct and has been proposed to be responsible for the symptoms due to obstruction of the bile and pancreatic ducts [2,3].

If a radiographic investigation shows a filling defect in the pancreaticobiliary system, a protein plug should usually be suspected and the simultaneous occurrence of symptoms may be explained by the consequent disturbance of biliary pancreatic secretory flow. This finding may be supported by analysis at the time of surgery [3]. A common channel protein plug complicating choledochal cyst is found in 10%–40% of cases, however, the

association of a pancreatic protein plug is considered rare [3,4]. The mechanism of protein plug formation in choledochal cysts has been gradually elucidated [5], but does not still become clear. The protein plug may act as a ball valve and produce a transient and abrupt elevation in intraluminal pressure in both the bile and pancreatic duct [6].

Kamisawa and co-workers reported that a filling defect disappeared spontaneously in 50% of patients [7]. However, if protein plugs remain incarcerated or persist until the time of surgery, a preoperative or intraoperative maneuver for protein plug extraction is chosen. Although the treatment of protein plugs located in the pancreatic duct has not been discussed well, the representative English language literatures on management of pancreaticobiliary protein plugs associated with choledochal cyst in children are displayed in Table 1. Preoperatively, endoscopic short-tube stenting was effective when ERCP showed the common channel protein plug impaction in patients with persistent or worsening symptoms [8] and endoscopic protein plug extraction or irrigation for the washout in the common channel is adequate and effective for the management of small protein plugs as well [9,10]. On the other

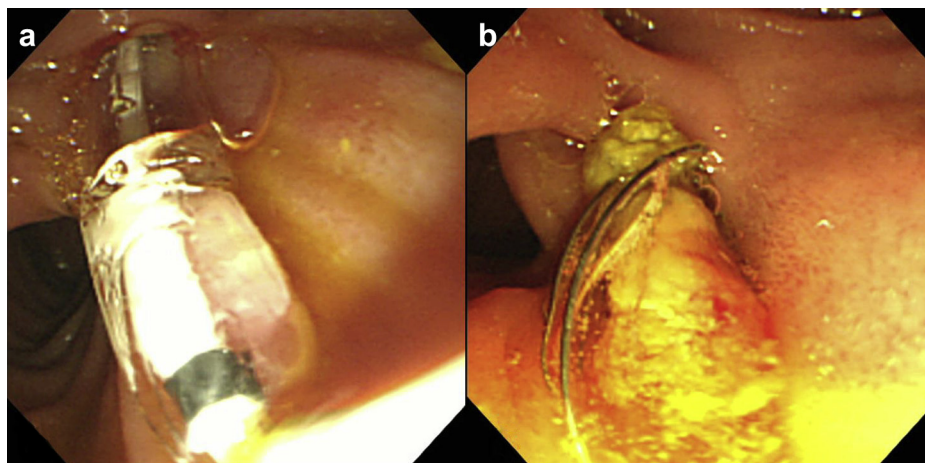


Fig. 2. a) Endoscopic sphincteroplasty and b) protein plug extraction. The protein plug was stagnant despite the washout-effect by contrast medium. Endoscopic sphincteroplasty was followed by extraction of the protein plug with a basket lithotripter. The plug was crushed but extracted as a mass via the papilla of Vater.

Table 1

Characteristics and management of pancreaticobiliary protein plugs (PP) associated with choledochal cyst (CC): analysis of literature written in English.

Author	Year	Duration (years)	CC case	PP case (% of CC)	Age, mean (range)	Location of PP	Preoperative maneuver		Intraoperative maneuver (n)	Miscellaneous (n)
							Therapeutic ERCP (n)	Others (n)		
Tsuchiya et al. [8]	2013	5.5	55	12 (21.8 %)	2.9 years (median)	Common channel narrow segment of CC	Short-tube stenting (11) Crushing PP with guide wire (1)			Unsuccessful stenting (1)
Miyano et al. [11]	2011	1.2	12	7 (58.3 %)	N/A	Common channel			Lap-assisted clearance using endoscope via the end of CC (7)	
Diao et al. [12]	2010	8	218	34 (15.6 %)	4.98 years (1.0–16.5 years)	Common channel			Lap-assisted clearance using endoscope/catheter via the end of CC (34)	
Terui et al. [9]	2008	11	37	4 (10.8 %)	3.25 years (1–8 years)	Common channel	Sphincterotomy with basket extraction (4)	PTCD (1)		Failed therapeutic ERCP (1)
Houben et al. [10]	2007	7	42	7 (16.7 %)	3 years (2–12 years)	N/A	Short-tube stenting (2) Sphincterotomy with basket extraction (2) Basket extraction with short-tube stenting (2)	PTCD (1)		Hemorrhage at sphincterotomy (1)
Yamataka et al. [4]	2000	12	67	8 (11.9 %)	(3–7 years)	Pancreatic duct common channel			Irrigation using endoscope via the end of the CC (4) via the papilla of Vater (1) via the minor papilla (1) Incision of the pancreatic duct (2)	Postoperative pancreatitis (1)
Ando et al. [13]	1998	27	126	43 (34.1 %)	4.9 years (4 months–14 years)	Common channel		PTCD/T-tube (6)	Removal using blunt spoon via the end of CC (4) Removal with the residual cyst (1) Irrigation via the end of CC (1) Incision of the pancreatic duct (4)	Spontaneous disappearance (26) Therapeutic ERCP after cyst excision (1)

hand, several authors have reported that protein plugs should be flushed into the duodenum by irrigation through a thin tube or biliary scope placed in the common channel through the distal end of the choledochal cyst at the time of cyst excision [4,7,11–13].

In our institution, ERCP has become an accepted modality in pediatric patients, and preoperative ERCP is indicated for patients who are more than three years old and have a body weight of more than 10–15 kg. A standard medium-diameter (11.0–12.0 mm) duodenoscope with a 3.2-mm operating channel may be used in most children older than 2–3 years of age. The channel accommodates medium-sized stents and the passage of dilating catheters up to 7 Fr. Most endoscopic therapies used in adult patients have been performed in children using standard adult duodenoscopes [14]. On the other hand, a duodenoscope with a smaller diameter has been developed for children: it is only 7.5 mm wide and has a 2.0-mm diameter operating channel that accepts a 5 Fr or smaller cannula (Olympus PJF-160). This instrument is designed for neonates and infants who weigh less than 10 kg or who are less than 2 years of age [15]. However, this duodenoscope generally permits diagnostic ERCP and limited therapeutic ERCP only with adequate suction and the use of 5 Fr-wide accessories.

Although ERCP has been repeatedly shown to be feasible in children and infants in small case series [16], the small number of pediatric ERCP cases may not provide sufficient experience for pediatricians and pediatric surgeons to reach minimum training thresholds or to sustain the level of competence needed for therapeutic ERCP. As a result, most ERCP procedures in children are performed by adult gastroenterologists who perform a high volume of procedures. Within our medical center, a partnership has formed between pediatric surgeons and adult gastroenterologists which combines the necessary cognitive and technical skills to provide optimal care for pediatric patients. Complications of ERCP include pancreatitis, infection, hemorrhage, etc. and the rate of complications in children has been reported to be under 10% [17]. The most common complication is pancreatitis, which usually resolves in several days.

In this case, the symptoms resolved with conservative therapy, however, the protein plug was still deeply impacted in the main pancreatic duct, and therefore we considered that the protein plug should be removed from the papilla of Vater and planned to extract it endoscopically. The plug could not be moved or eliminated by irrigation with contrast medium. We needed to use a basket extractor to take it out as a lump or crush it within the duct. Fortunately, we could insert the basket extractor even though the plug was impacted, but endoscopic sphincteroplasty was required for plug extraction. We safely performed ERCP with plug extraction and the patient did not develop post-ERCP pancreatitis.

3. Conclusion

ERCP played an important role as a bridge therapy to surgery in our case. In cases in which a huge protein plug is deeply impacted in the main pancreatic duct, therapeutic ERCP may be useful for preparing for choledochal cyst excision with hepaticojejunostomy.

Further refinements in the design of pediatric duodenoscopes and proper equipment will likely be needed to minimize the risk of ERCP-related complications.

Conflict of interest

The authors have no conflicts of interest to report.

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